An unusual case of Multiple Inflammatory Myofibroblastic Tumors of the lung.

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ABSTRACT: Inflammatory myofibroblastic tumor (IMT) of the lung is a rare, usually solitary lesion that is considered nowadays as a true neoplasm with unpredictable clinical course. Herein we describe a case of multiple, bilateral IMTs of the lung in a 70 year-old asymptomatic woman, where diagnosis was established by CT guided core biopsy. To our knowledge only 10 cases of bilateral IMTs have been reported in a few case series and in only 2 cases of pulmonary IMTs, CT guided biopsy could establish the correct diagnosis.

Key Words: Inflammatory myofibroblastic tumor, Inflammatory pseudotumor, Lung, Computed tomography.

INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) of the lung is a rare, usually solitary lesion. IMTs are considered as benign true tumors, though some authors regard these lesions as low-grade sarcomas. The behavior of these lesions is unpredictable and complete resection should be considered as the treatment of choice[1,2].

Herein we describe a case of multiple, bilateral IMTs of the lung in a 70 year-old asymptomatic woman, where diagnosis was established by CT guided core biopsy. To our knowledge, only 10 cases of bilateral IMTs have been reported in 6 case series[3-8], and only 2 cases of pulmonary IMT were correctly diagnosed by CT guided biopsy[1].

CASE REPORT

A 70 year-old asymptomatic woman with incidentally found bilateral lung lesions during pre-operative control was referred to our department for CT (Figure 1A). Physical examination was unremarkable. Contrast-enhanced CT demonstrated 3 lung lesions· 1 lesion of 11 mm in the right upper lobe, 1 lesion of 41mm in the lingula and 1 lesion of 55 mm in the left lower lobe. The rest lung parenchyma was normal and no mediastinal lymph nodes were demonstrated (Figure 1A-D).

Due to suspicion of metastatic disease, the patient was referred for CT guided percutaneous core biopsy. The lesion in the lingula was considered to have the easiest access for biopsy and 4 tissue shamples were obtained with an 18 Gauge cutting needle using the coaxial technique (Temno;Evolution;Cardinal Health;Orlando) (Figure 1E). Pathological examination was performed by two pathologists (I.E, A.F) and demonstrated bundles of collagen fibers and the characteristic spindle-cells, which at immunohistochemistry were positive in vimentin and smooth muscle actin (SMA) and negative in CD34 and BCL-2. There were also areas of chronic inflammatory infiltrations with the presence of lymphocytes and plasma cells. Lung parenchyma with inflammation was also found adjacent to the lesion in some tissue specimens (Figure 1G-I). These findings were consistent with lung IMT. A second biopsy of the lesion in the left lower lobe

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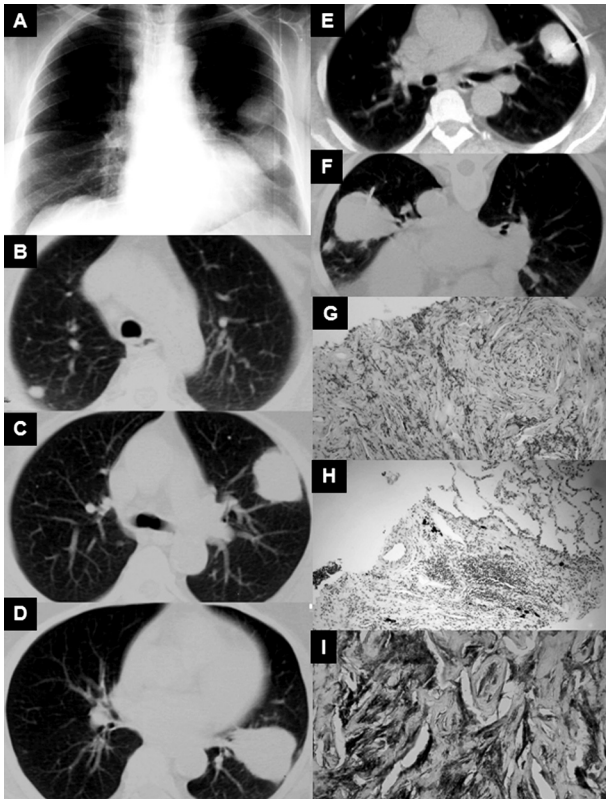

Figure 1. A. Preoperative chest x-ray, showing bilateral lung lesions, one small in the right upper lobe and two larger in the left lung.
B-D. Chest CT in lung window demonstrating the three lung lesions.
E-F. CT guided biopsy of the lesions.
G. Hyalinized connective tissue with scattered chronic inflammatory infiltrations. (Hematoxylin & Eosin X200).
H. Lung parenchyma adjacent to the lesion. (Hematoxylin & Eosin X100).
I. Smooth muscle actin positive spindle cells. (SMA x400).

was performed some weeks later in order to confirm the nature of the lesion and to exclude coexistence of malignancy. Performing a posterior approach in order to avoid transversing the interlobar fissure, 4 tissue shamples were obtained with the same technique as described above (Figure 1F). Specimens were examined by the same pathologists and demonstrated the same histological findings with the first lesion. The lesion in the right upper lobe was considered as IMT without biopsy.

The patient after being informed about the disease chose not to be operated or receive any treatment since she was completely asymptomatic, and she was advised to have a chest CT scan every 6 months. One year after the initial diagnosis, the lesions remained unchanged in follow up CT, and the patient was asymptomatic.

DISCUSSION

IMT of the lung is a rare and not well known disease, accounting for 0,7-1 % of all thoracic tumors, while in children under 16 years, IMT is the most common primary tumor[1,9-11]. Other terms used for IMT include inflammatory pseudotumor, plasma cell granuloma, xanthogranuloma and fibrous histiocytoma[1,12,13].

IMTs of the lung can be found at any age group including childhood, but are more often found in patients younger than 40 years old .There is no sex predilection and in about half of the cases patients are asymptomatic and lesions are incidental findings in chest x-rays[1,3,6,10,12]. Symptomatic lung IMTs may cause dyspnea, cough, hemoptysis, chest pain, fever and general weakness[5]. Lung IMTs are usually solitary peripheral lung lesions and only 10 cases of multiple IMT‘s have been mentioned in 6 case series[3-8]. IMTs can be also found in multiple other anatomic locations including bladder, spleen, breast, pancreas, liver, colon, spermatic cord, prostate, peripheral nerves, soft tissue and orbit[2].

It is believed that IMTs of the lung are the result of an unregulated response of inflammatory cells to tissue damage of chronic inflammation, related to pulmonary infection[3,10]. No genetic or environmental factors predispose for lung IMTs[1]. Although IMTs of the lung have been considered to be benign lesions, they have an unpredictable and variable clinical course, with local invasion, local postoperative recurrences and distant metastases in some cases[1,3,14]. Chromosomal abnormalities detected in the younger age group IMTs suggest that lung IMT might be a true neoplasm rather than a purely inflammatory or reactive lesion[14,15]. One-third of cases show a 2p23 rearrangement involving anaplastic lymphoma kinase (ALK), often fused with tropomyosin genes TPM3 and TPM4 and the clarithrin heavy chain gene (CLTC)[16]. That is the reason why some authors consider these tumors as low-grade malignancies and propose a more aggressive treatment[1,12,17].

Cerfolio et al. divided IMTs into noninvasive and invasive. The invasive form is more often in pediatric patients (about 20%) who are at high risk for locoregional extension[10]. Lung IMTs may invade the mediastinum, the chest wall, the contralateral lung, large vascular structures, the left ventricle, the vertebra, the diaphragm, the thoracic inlet, the main bronchi and the carina[1,18]. In such cases, complex debulking operations may be needed. Histologically lung IMT's are characterized by the presence of myofibroblasts (spindle cells) with varying infiltrations of chronic inflammation and plasma cells[12,19].

There are not specific guidelines about treatment of IMTs since these tumors are rare and clinical course varies. Most authors propose a more aggressive treatment, with complete resection when possible, since patients with IMT's are usually young and these tumors often have malignant behavior[1]. Chemotherapy, steroid therapy and radiation therapy have been used by some authors with variable results[1].

CT guided core biopsy or fine needle aspiration (FNA) have been used by some authors for the diagnosis of IMTs with disappointing results[1,3,6,7,11,12]. CT guided FNA has been described as non diagnostic for lung IMT's[3,12]. CT guided core biopsy has also be described as non diagnostic in most cases[6,7,11]. Only Fabre et al. managed preoperative diagnosis of 2 lung IMTs by CT guided biopsy[1]. In our case, both biopsies that were studied by two pathologists were diagnostic for lung IMT. It seems that adequate tissue specimen is necessary for the diagnosis of lung IMTs. When small tissue specimen is taken by core biopsy and in samples from FNA, the mixture of chronic inflammatory cells and fibroblastic proliferation cannot lead to a definite and safe diagnosis because it is also found in a variety of other inflammatory and malignant lesions[11,12].

Abbreviations: *Inflammatory myofibroblastic tumor (IMT), computed tomography (CT), fine needle aspiration (FNA).*

Ασυνήθης περίπτωση πολλαπλών φλεγμονωδών μυοϊνωβλαστικών όγκων πνεύμονα.

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ΠΕΡΙΛΗΨΗ: Οι φλεγμονώδεις μυοϊνωβλαστικοί όγκοι (ΦΜΟ) του πνεύμονα είναι σπάνιες, συνήθως μονήρεις πνευμονικές βλάβες οι οποίες σήμερα θεωρούνται ως αληθή νεοπλάσματα με απρόβλεπτη εξέλιξη. Περιγράφουμε μια περίπτωση πολλαπλών, αμφοτερόπλευρων ΦΜΟ του πνεύμονα σε μια 70χρονη ασυμπτωματική γυναίκα όπου η διάγνωση τέθηκε με κατευθυνόμενη βιοψία με τη βοήθεια του αξονικού τομογράφου. Εξ' όσων γνωρίζουμε, λιγότερες από 10 περιπτώσεις αμφοτερόπλευρων ΦΜΟ έχουν αναφερθεί και μόλις σε 2 περιπτώσεις η κατευθυνόμενη βιοψία έθεσε την ορθή διάγνωση.

Λέξεις Κλειδιά: Φλεγμονώδης μυοϊνωβλαστικός όγκος, Φλεγμονώδης ψευδοόγκος, Αξονική τομογραφία.

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